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# Risk of Aborted Cardiac Arrest or Sudden Cardiac Death During Adolescence in the Long-QT Syndrome

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**Context** Analysis of predictors of cardiac events in hereditary long-QT syndrome (LQTS) has primarily considered syncope as the predominant end point. Risk factors specific for aborted cardiac arrest and sudden cardiac death have not been investigated.

**Objective** To identify risk factors associated with aborted cardiac arrest and sudden cardiac death during adolescence in patients with clinically suspected LQTS.

**Design, Setting, and Participants** The study involved 2772 participants from the International Long QT Syndrome Registry who were alive at age 10 years and were followed up during adolescence until age 20 years. The registry enrollment began in 1979 at 5 cardiology centers in the United States and Europe.

**Main Outcome Measures** Aborted cardiac arrest or LQTS-related sudden cardiac death; follow-up ended on February 15, 2005.

**Results** There were 81 patients who experienced aborted cardiac arrest and 45 who had sudden cardiac death; 9 of the 81 patients who had an aborted cardiac arrest event experienced subsequent sudden cardiac death. Significant independent predictors of aborted cardiac arrest or sudden cardiac death during adolescence included recent syncope, QTc interval, and sex. Compared with those with no syncopal events in the last 10 years, patients with 1 or 2 or more episodes of syncope 2 to 10 years ago (but none in the last 2 years) had an adjusted hazard ratio (HR) of 2.7; (95% confidence interval [CI], 1.3-5.7; $P<.01$) and an adjusted HR of 5.8 (95% CI, 3.6-9.4; $P<.001$), respectively, for life-threatening events; those with 1 syncopal episodes in the last 2 years had an adjusted HR of 11.7 (95% CI, 7.0-19.5; $P<.001$) and those with 2 or more syncopal episodes in the last 2 years had an adjusted HR of 18.1 (95% CI, 10.4-31.2; $P<.001$). Irrespective of events occurring more than 2 years ago, QTc of 530 ms or longer was associated with increased risk (adjusted HR, 2.3; 95% CI, 1.6-3.3; $P<.001$) compared with those having a shorter QTc. Males between the ages of 10 and 12 years had higher risk than females (HR, 4.0; 95% CI, 1.8-9.2; $P = .001$), but there was no significant risk difference between males and females between the ages of 13 and 20 years. Among individuals with syncope in the past 2 years, β-blocker therapy was associated with a 64% reduced risk (HR, 0.36; 95% CI, 0.18-0.72; $P<.01$).

**Conclusions** In LQTS, the timing and frequency of syncope, QTc prolongation, and sex were predictive of risk for aborted cardiac arrest and sudden cardiac death during adolescence. Among patients with recent syncope, β-blocker treatment was associated with reduced risk.

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